

Letters to the Editor

The medullary carcinoma of the pancreas. A relative new entity

Key words: Medullary carcinoma. Pancreatic adenocarcinoma.

Dear Editor,

The medullary carcinoma of the pancreas is a relatively new variety of adenocarcinoma of pancreas. It has been recently described and there are very few clinical reports in the literature. Wilentz et al. (1) reported about their experience with this new entity and they analyzed the genetic aspects in this variant of pancreatic adenocarcinoma. The prognosis is better than infiltrating ductal adenocarcinoma of the pancreas, and there are not special recommendations about treatment. In general, the treatment of this tumor is similar to infiltrating ductal adenocarcinoma of the pancreas (surgery, chemotherapy and radiotherapy).

Case report

We report the case of a 59-year-old man with diabetes type 2 (in treatment with insulin), arterial hypertension, hypercholesterolemia, hypothyroidism, chronic obstructive pulmonary disease, auricular fibrillation and myocardial infarction in 1998. The patient entered in the hospital owing to the fact that it presented elevation of the level of bilirubin in blood. The abdominal ultrasound was normal and abdomen magnetic resonance image (MRI) showed a mass in the body of the pancreas without vascular permeation. The team of surgeons decided to operate him. The technique used was the procedure of Whipple.

The histological examination showed one pancreatic carcinoma with necrotic areas, expanding pattern of invasion, and syncytial growth, and was diagnosed as a medullary carcinoma of pancreas. Ten of twenty-four lymph nodes were affected, and margins of resection were affected. Tumor cells were positive for citoqueratins AE1/AE3 and 7. These cells had small positive EMA, and were negative for citoqueratin 20, CEA, neuroendocrine markers, S100, CD45. k-ras was wild type.

Two months after the surgery the performance status of patient was 3. New analysis performed showed a relapse of the tumor located in lymph nodes and in the peritoneal liquid.

The evolution of the patient was unfavorable, with a worsening of the symptoms, and was not possible to start any treatment with chemotherapy.

Finally, the patient died due to tumor progression, five months after the diagnosis.

Discussion

Sometimes in our clinical practice we find with strange tumors variants. This is one example. There are not specific guidelines for the treatment of these tumors and if we must to follow treatment recommendations for the pancreatic adenocarcinoma.

The medullary carcinoma of the pancreas is a rare variant of adenocarcinoma pancreatic, described for the first time in 1998 (2). The medullary phenotype is characterized by poor differentiation, and expanding pattern of invasion, and syncytial growth.

It is important to know that medullary carcinoma has a special genetic profile and it is different from conventional pancreatic ductal adenocarcinomas. Approximately 69% of these tumors have wild-type k-ras genes and 22% of these tumors have microsatellite instability (MSI) (1).

The MSI is a very important difference with typical adenocarcinoma, in which is very unusual to have MSI. This difference is significant because we can use this tumor to identify patients with susceptibility to cancer.

Banville et al. (3) report a case of a man with colorectal carcinoma who developed pancreatic medullary carcinoma. The tumor showed MSI and loss of expression of the mismatch repair proteins MSH2 and MSH6, and finally was diagnosed of hereditary nonpolyposis colorectal cancer (HNPCC).

Lynch et al. in 1985 (4) reported pancreatic cancer in some HNPCC kindred suggesting that pancreatic carcinoma may be integral to the HNPCC phenotype. The significance of this difference between typical adenocarcinoma and medullary carcinoma is important because suggest that medullary features in pancreatic carcinoma can identify patient with one genetic predisposition for cancer. Finally, Wilentz et al. (1) report a patient with pancreatic cancer and latent Epstein-Barr virus. This medullary carcinoma had lymphoepithelioma-like features, with infiltration of the neoplasm by large numbers of lymphocytes. It is a new feature to investigate in this type of tumors.

In conclusion, it is very important to identify this type of tumor due to the relationship between them and genetic cancer predisposition.

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